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DISSECTING ANEURYSM OF THE AORTA

Since the publication of Shennan's excellent monograph on dissecting aneurysm in 1934, there has been a marked increase in the frequency with which this condition has been correctly diagnosed. The possibility of diagnosis depends largely upon the physician's familiarity with the pathological and clinical features, and to only a limited extent upon diagnostic aids. Outstanding clinical features are illustrated by three cases summarized below, detailed reports of which have been made elsewhere, in which the antemortem diagnosis was correctly made.

Case 1.—A clergyman, 45 years of age, had palpitation, mild dyspnoea, fatigue, a brief episode of auricular fibrillation, and some retrosternal discomfort extending to the left arm. After three months of this he collapsed suddenly, in mild shock, without chest pain but with signs of aortic insufficiency, and flaccid paralysis of both legs with sensory disturbances, especially on the right. There was fever, leukocytosis and mild anemia. Later there was transient hematuria from the right kidney. Towards the last there were brief attacks of fairly intense cyanosis, especially during sleep. The circulation in the legs became markedly enfeebled, especially on the right. Death occurred forty days after the onset. Postmortem examination showed an intimal rent almost completely encircling the aorta, about an inch above the valve. From this point dissection had extended proximally and pointed backwards and to the right within the pericardium, compressing the right auricle. The dissection also extended distally throughout the entire length of the aorta, stopping at the bifurcation. The terminal portion was filled with thrombus and caused marked obstruction of the orifices of the iliac arteries. On the right the third to eleventh, inclusive, intercostal arteries, were severed by the aneurysm. The subcostal and lumbar arteries were severed bilaterally. The renal arteries were obstructed, especially the right.

Case 2.—A retired postman, 68 years of age, with a preexisting hypertension and nephritis, had a sudden onset of intense pain within the chest, with shock. The pain extended to the back and to the epigastric region, but did not radiate to the arms. There was no actual dyspnoea and little weakness, but fever ranging to 102° F., and mild retrosternal pain persisted to the end. There were no electrocardiographic signs indicative of coronary occlusion. Death occurred suddenly about six weeks after the onset. At postmortem an intimal rent was found almost completely encircling the aorta in the descending portion of the arch. Dissection had occurred proximally about three centimeters and distally about eighteen centimeters. The terminal portion of the aneurysm was filled with thrombus. In the central portion rupture had occurred laterally into the left pleural cavity, which was filled with blood.

Case 3.—A male executive, 43 years of age, with a preexisting hypertension, had a sudden onset of constricting pain behind the upper sternum which quickly passed downward to the abdomen, through to the back, and down the spine to the lumbar region. Very quickly thereafter numbness of the left leg was noted and an inability to move the left foot. The pain subsided within twenty-four hours. There was low-grade fever and leukocytosis. Variable and bizarre neurologic manifestations in the left leg persisted for about two weeks. Left hematuria developed about this time. Recovery from the acute manifestations was apparently complete except for recurrence of the hematuria. Death occurred twenty-seven months after the onset, with signs of subarachnoid hemorrhage. Postmortem examination showed a dissecting aneurysm which began just distal to the orifice of the left subclavian, and passed downward and a little to the left within the posterior wall of the aorta, and extended into the left iliac artery as far as this vessel was traced. The true aortic channel became obliterated above the bifurcation and the right iliac was supplied by rupture of the aneurysm. All the intercostal and lumbar branches on the left were ruptured, and the aneurysm had penetrated the left renal and the superior mesenteric arteries.

There are two areas of predilection for the initial rupture; the ascending portion of the aorta shortly above the valve, and the isthmus or site of the ligamentum arteriosum. The former may be subjected to mechanical stress by diastolic recoil following closure of the aortic valve. The latter may be rendered weak by defects resulting from obliteration of the ductus arteriosus. An atheromatus ulcer is infrequently the site of rupture. Medial degeneration characterized by round cell infiltration, the presence of lipoid giant cells, fragmentation of elastic tissue and hyalinization of fibrous tissue have been present in many cases. These changes are focal and tend to become vacuolated or cystic (medionecrosis of Erd-

heim.) The vasa vasorum may be found obliterated or showing inflammatory or sclerotic changes, and the suggestion has been made that intramural hematomata, as a result of disease of the vasa vasorum, may precede intimal rupture. The course of dissection lies between the layers of the media. Small branches of the aorta lying in the path of the aneurysm are at first left connected with the true aortic channel by a slender tube of intima, which is drawn out and compressed and finally severed as the size of the aneurysm increases. Larger branches may be involved by direct extension of the aneurysm within their walls. Extensive aneurysms usually pass down the posterior wall of the aorta and thus interrupt several of the intercostal and lumbar arteries. The interference with blood supply of the spinal cord so produced gives rise to degenerative changes within the cord, and to the patchy and bizarre neurological findings illustrated in the cases described. Larger vessels such as the renal or mesenteric arteries may be penetrated by the aneurysm with resultant symptoms such as anuria or hematuria, abdominal distention, ileus or melena. Penetration or compression of major branches may give rise to pulse and pressure changes in the extremities, or to tenderness along the involved arteries. In about four cases of five, the duration of life after the formation of a dissecting aneurysm will be measured in hours or days. When the rent in the intima is situated shortly above the aortic valve, dissection often proceeds proximally and may rupture into the pericardium. Inspection of the illustrations in published case reports seem to indicate a frequent point of rupture external to the right posterior sinus of valsalva, corresponding to the point of greatest aneurysmal distention in Case 1 above. Ruptures into the mediastinum and into the pleural cavities are also frequent. External rupture lower down may give rise to extreme extravasation of blood before death. Rupture of the aneurysm back into the main channel of the aorta may permit recovery to occur with the formation of a so-called "double barreled" aorta. In such cases life might presumably go on for years, and death occur from an extraneous cause as in Case 3 above. An aorta of this type has been found at autopsy in cases without history of previous severe illness, and in which no idea of the probable duration could be obtained.

In cases of dissecting aneurysm of the aorta, antecedent hypertension is frequent and syphilis is infrequent. Most cases have occurred in males between forty and sixty years of age. The onset is usually sudden, with agonizing pain of a rending character, the location of which varies with the site of the aneurysm, but which characteristically exhibits a migratory tendency. It may begin anteriorly in the precordial region or in the epigastrium, pass to the neck or jaw, later to the back, the flanks and the legs, or any of these areas may be involved. Radiation of pain to the arms is perhaps less common than in myocardial infarction. The onset is also usually attended by pallor, perspiration and prostration, and often by loss of consciousness. The latter is especially likely if the carotid arteries are involved by the dissection, in which event hemiplegia or other central nervous manifestations may supervene. When intimal rupture occurs in the descending thoracic or in the abdominal aorta, confusion with renal or intra-abdominal disease is most likely. External rupture in this situation may result in extensive extravasations of blood which, if detectable externally, may be thought to require surgical exploration, or, forming slowly, may give rise to jaundice from absorption of blood pigment. Pain may be remarkably absent even when the dissection is extensive, as in Case 1 above. The diagnosis of coronary occlusion, particularly in the absence of characteristic eletrocardiographic findings, should not be finally made without considering other possibilities, including dissecting aneurysm. Sudden onset of severe pain and prostration are common to both, but the pain of the latter is even more abrupt and even less amenable to relief, and in patients that survive, it tends to recur over longer periods. The tendency of the pain of dissecting aneurysm to migrate from front to back and from above downwards, should also be kept in mind. A history of anginal syndrome speaks for coronary thrombosis. The development of neurological signs from ischemia of the spinal cord can greatly facilitate the diagnosis of dissecting aneurysm. Search should be made for neurologic changes either motor or sensory or both: perhaps transitory, but characteristically segmental in distribution. It is possible that the symptoms and signs of peripheral neuritis may at times arise from ischemia of nerve trunks. These manifestations are to be distinguished from symtoms due to occlusion of the arteries of the extremities, which may be present at the same time or later. The occurrence of gross hematuria or of anuria, indicating involvement of the renal arteries, may also be an important clue. Hematuria will have enhanced significance in this connection if it can be shown to be unilateral in origin, and to come from the side on which the neural lesions are most extensive. It is only in healed cases that Roentgen examination may be expected to contribute materially to the confirmation of the diagnosis.

There is unfortunately little which may be done in treatment. Absolute rest should be enforced with opiates and barbiturates. The blood pressure should be lowered with nitrites. Venesection has been advised, but the essential reduction in pressure should usually be obtainable with the vasodilators. Operative intervention to reunite the cavity of the aneurysm with the main channel of the aorta just above the bifurcation has been conceived and in one instance attempted, but a successful outcome in such a venture must necessarily require a fine combination of circumstance, diagnostic acumen, and surgical skill. Patients who have recovered should of course have their activity markedly restricted, yet the enjoyment of life and even the opportunity of earning a livelihood need not necessarily be denied them.

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